California MEDICINE

OFFICIAL JOURNAL OF THE CALIFORNIA MEDICAL ASSOCIATION
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Volume 87 DECEMBER 1957 Number 6

Diagnosis of Hemorrhagic Diseases

Evaluation of Procedures

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• Careful and complete history and physical examination are essential in the diagnosis of diseases characterized by hemorrhage and are more reliable than laboratory tests for the prediction of the tendency to bleed at the time of surgical operation. Specific questions should be asked about bleeding from various anatomical sites, allergic manifestations, diet, and exposure to poisons and chemical substances. Because hemorrhagic diseases may be hereditary, it is necessary to obtain a family history. If there is a personal or family history of abnormal bleeding, the examiner should obtain exact details about the events immediately preceding the bleeding episodes, the character and duration of the hemorrhage and the response to therapy.

The history is particularly important in the case of patients who are to have operation. Failure to obtain a history of past bleeding episodes may be catastrophic. In patients with bleeding tendencies who require operation, the history is valuable in predicting the severity of the bleeding.

Physical examination including examination of retina, breast and pelvic organs will often detect evidence of lesions that are helpful in the diagnosis of the primary diseases of which the hemorrhage is a manifestation. The location, distribution, character and number of hemorrhagic lesions are of value in diagnosis and in prognosis.

PART I

History, Physical Examination

WHEN A PATIENT seeks medical care because of ecchymoses, petechiae, spontaneous bleeding from mucous surfaces or abnormal bleeding following trauma, the procedure to be followed is the same as it would be for any other patient. Hemorrhagic phenomena are manifestations of disease of varied etiologic factors. The primary disease may involve any organ of the body or the body as a whole. It is therefore necessary to utilize all of the diagnostic tools, including history, physical examination and screening laboratory procedures as well as special tests. It is the purpose of this paper to discuss the role that the history and the physical examination play in the diagnosis of diseases characterized by hemorrhage.

HISTORY

The "present illness" of a patient with hemorrhagic phenomena should start at the time of birth, since diseases characterized by abnormal bleeding may be hereditary and since the defect is likely to become manifest in infancy or in early childhood. All persons are continually subjected to tests of trauma and often react in characteristic ways to injuries. Persons in normal environments bump into objects, are cut, scratched, bitten and spanked. Sneezing, coughing, vomiting, defecation, shaving, cleaning the teeth, menstruation, the wearing of tight clothing and assumption of the upright posi-

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Guest Speaker's Address: Presented before the Section on Pathology and Bacteriology at the 86th Annual Session of the California Medical Association, Los Angeles, April 30 to May 1, 1957.

Part I of a two-part article.

tion produce stresses on the vascular system. Inquiry should be made concerning accidents, surgical operations, extraction of teeth and childbirth. If a patient is one or more years of age and his system review reveals no tendency to bruise easily and no abnormality in hemostasis following trauma, and particularly if there has been no abnormal bleeding following cuts, fractures or the extraction of teeth, one is safe in assuming that hereditary hemorrhagic disease does not exist.

Many people who bruise easily or who bleed abnormally have a history of allergic sensitivity; hence, it is important to ask specific questions about eczema, urticaria, erythema, hay fever, asthma and sensitivity to foods and drugs.

The history should also include inquiry about exposure to chemicals, sprays, insecticides, cleaning solutions and solvents in the home and at work. A list should be made of all drugs taken by mouth or parenterally. If the patient has been taking medicines, the timing of the hemorrhagic state in relation to the taking of a given drug is important. Many patients do not consider that a given substance is a drug unless it is prescribed by a physician. It is therefore necessary to ask about sleeping pills, hair rinsing fluids, cosmetics and deodorants, contraceptives, douches, powders, salves, lotions, headache remedies, cathartics, mouthwashes, gargles and hair dyes.

The history should also include questions concerning the diet. A deficiency in vitamin C may be the cause of scurvy which is characterized by gum bleeding, ecchymosis and subperiosteal hemorrhage. Deficiencies in protein, calcium, iron and vitamins are seldom the primary cause of bleeding, but often are factors in exaggeration of the hemorrhagic state.

It is important to inquire carefully about events preceding the last hemorrhagic episode and particularly to inquire about "colds," "sore throat" and "fever." Patients with thrombocytopenic purpura often give a history of infection of the respiratory tract, exanthema or other febrile diseases occurring one or two weeks preceding the onset of hemorrhagic phenomena.

It is particularly important to obtain a past and present history of hemorrhagic phenomena in patients admitted to surgical services. Failure to obtain a careful history may lead to serious consequences as exemplified by the following cases.

CASE 1. A boy was admitted with a large fluctuant hematoma of the thigh, which appeared following a minor fall. No inquiry was made concerning his past history. The hematoma was lanced and drained, following which there was massive bleeding, which could not be readily controlled and which necessitated multiple transfusions. The history obtained later revealed that the patient previously had had many similar extravasations of blood into the soft tissues, that he had several ankylosed joints, that he bled severely following minor cuts and that he had siblings who were "easy bleeders."

CASE 2. A young man was admitted to the neurosurgery service because of head injury received in an automobile accident. At the time the patient was admitted he was mentally clear and all vital signs were normal. Several hours later he lapsed into coma and died. The family later volunteered the information that the patient had always bled easily following trauma and that he had been diagnosed as having "hemophilia." If the history of previous bleeding had been obtained from the patient and if transfusions had been instituted at the time of admission, the catastrophe might have been averted.

Patients with defects in coagulation mechanisms develop conditions requiring surgical procedures as do patients with normal hemostatic and coagulation mechanisms. In such persons the history is of great value in predicting the degree of difficulty to be expected and the calculated risk that the patient must assume. For example:

CASE 3. A patient with capillary purpura due to hereditary capillary fragility (pseudohemophilia) bled excessively at each menstrual period. In spite of the excessive blood loss, the patient had not become anemic, she had borne two children without requiring transfusions and had had several teeth extracted with some difficulty in hemostasis, but with no massive bleeding. The bleeding time was only slightly prolonged. In view of the history of minor bleeding following trauma, it was the opinion of the consultant that hysterectomy could be performed with reasonable safety. There was only moderate difficulty in hemostasis at the time of operation and no postoperative bleeding.

CASE 4. A patient with massive ecchymoses of legs and arms, thought to be due to sensitivity to drugs, did not bleed abnormally at the time of biopsy of a cervical lymph node. On the basis of this history and in spite of the positive physical signs, it was recommended that indicated pulmonary operation was possible. Lobectomy was accomplished without unusual loss of blood.

CASE 5. A patient with meningioma gave a history of extravasations of blood into muscles, causing disability and severe bleeding following trauma, and necessitating transfusions. The patient assumed the grave risk of hemorrhage rather than choosing to die of brain tumor. The operation could not be completed, for the bleeding was so excessive at the incision line that the skull flap could not be exposed.

If the patient gives a history of bleeding, the physician should obtain details about the timing of the hemorrhage in relation to the trauma, severity of the hemorrhage and the duration and response to therapy. Patients tend to overemphasize bleeding and many patients think that they are "easy bleeders" when they are not. Occasional epistaxis and

bruises are to be expected in every normal person. Bleeding is of a serious nature when it causes the patient to seek medical aid, to receive transfusions or to be admitted to the hospital. Bleeding following extraction of teeth is probably significant if the bleeding continues through the second and third day. Difficulty in hemostasis at the time of tonsillectomy is more significant than hemorrhage one week after the operation. Bleeding from gums following cleaning of teeth is significant if there are clots, blood on the pillow or hematemesis.

FAMILY HISTORY

The family history is essential in hemorrhagic diseases because there are many kinds of abnormality that are genetically determined. The simplest way to record the results of a family survey is by means of a genetic diagram with symbols to represent the type of abnormality noted in siblings, children, parents, grandparents, uncles and aunts.

The physician should not be content when a patient recalls that a given member of the family was a bleeder, but should obtain as much detailed information as possible concerning the nature of the bleeding. Without such information, erroneous conclusions could be drawn, Thus, Uncle Charlie bled to death, but his arm was cut off in a sawmill accident. Aunt Susie died at the time of labor from blood loss, but she had previously given birth to six children without difficulty. The cause of her death was placenta previa and afibrinogenemia.

The absence of history of known bleeders in a family does not exclude the possibility of hereditary disease, for the abnormality may not be expressed for several generations or may be so mild as to go unnoticed. Many patients do not have enough siblings or relatives to permit an hereditary trait to become manifest.

PHYSICAL EXAMINATION

The physical examination often reveals abnormalities that are of value in the diagnosis of hemorrhagic diseases. Ophthalmoscopic examination may reveal petechiae, which are helpful in the diagnosis of subacute bacterial endocarditis in a patient in whom the major signs may be intestinal bleeding. Retinal tubercles may aid in the establishment of the diagnosis of miliary tuberculosis that may be the cause of pancytopenia and thrombocytopenic purpura. Retinal hemorrhage, together with the finding of ecchymosis of the thigh and a positive reaction to a tourniquet test and a slightly prolonged bleeding time, may be of great value in the differential diagnosis of retinal thrombosis and systemic hemorrhagic disease. Telangiectatic lesions of the skin, nailbeds, lips or mucous membranes may be the key to unexplained bleeding from the nose, lungs, gastrointestinal or urinary tracts. Spider nevi suggest liver disease. A smooth tongue and loss of vibration sense may be a clue to the diagnosis of pernicious anemia, which is sometimes characterized by bleeding.

Examination of a breast may reveal a small carcinoma which in its metastatic spread may cause thrombocytopenia and uterine hemorrhage.

Jaundice is a warning signal and is an indication for withholding surgical operation until prothrombin activity and other tests have been evaluated.

Careful physical examination should always be done before surgical operation. Petechiae, ecchymosis, gingival bleeding and appearance of blood in the nares are warning signals. Nurses, aides and technologists should be alerted and instructed to call the attention of the physician to any sign of blood in sputum, vomitus or feces. Abnormal bleeding from the skin following hypodermic injections or needle puncture wounds, or recurrence of bleeding after preliminary cessation of bleeding may be of major significance, particularly if the patient has been admitted for surgical treatment.

The location and the distribution of the hemorrhagic lesions are of diagnostic importance. In senile purpura the ecchymoses are usually over the backs of the hands and over the shins and the tops of the feet. In capillary purpura due to allergic sensitivity (Schönlein-Henoch type, nonthrombocytopenic purpura), the petechiae are most prominent over the pressure points, such as the backs of the elbows, the buttocks and the knees. In this disease the hemorrhagic phenomena are often associated with swollen, tender and painful joints and sometimes with hematuria and melena. Petechiae due to thrombocytopenic purpura are most likely to occur over the lower legs and arms and in the mucous membranes. In fat emboli the fine, brownish petechiae are likely to be prominent over lateral chest wall and axilla. In erythema multiforme exudativum (Stevens-Johnson syndrome) the hemorrhagic lesions may be limited to the mouth and mucous membranes.

Petechiae limited to the face, neck and shoulders suggest vascular disease due to increased venous pressure such as occlusion of the superior vena cava or stasis of blood in the head due to breathholding or severe coughing. Hemorrhagic lesions of the abdomen, legs or arms may be due to tightly fitting or elastic garments.

Massive extravasation of blood with stocking and glove patterns of the extremities are characteristic of purpura fulminans. These are probably a severe form of allergic purpura in which there is vascular disease with occlusion of major blood vessels. In many of the severe infections there may be petechiae all over the body due to hemorrhagic emboli as is

seen in meningococcic septicemia. The rose spots of typhoid fever, the hemorrhagic extravasations in rickettsial disease and the hemorrhagic rashes seen in some patients with exanthematous diseases are examples of hemorrhagic phenomena which are manifestations of lesions in the vascular bed leading to the escape of erythrocytes.

The presence of hemorrhagic lesions that are atypical and are difficult to correlate with any clinical pattern should make one consider self-inflicted lesions (purpura factitia). A narcotic addict in order to obtain morphine complained of severe kidney colic and added blood to her urine and induced bleeding by needle puncture wounds in the skin in order to make the claims of hemorrhage more impressive. A patient desiring attention and sympathy took Dicumarol in order to produce hematuria and other hemorrhagic phenomena. Hemorrhagic lesions are most commonly inflicted on accessible areas, such as the forearms and thighs and are not present on the posterior surfaces which are more difficult to reach. The examiner should always closely inspect hemorrhagic lesions for tooth and fingernail marks and for puncture wounds in the center of them in order to make sure they are not self-inflicted.

Morphological characteristics of the skin lesions may be of value in diagnosis. Multiple petechiae which are of pinhead size are suggestive of thrombocytopenia or systemic vascular disease. Larger ecchymoses without petechiae suggest defect in the coagulation components. Hemorrhages into the muscles, joints or deep extravasations of blood along fascial planes suggest defects in plasma components such as occur in hemophilia, fibrinogenopenia and diseases caused by an increase in anticoagulants.

Large bruises of the superficial skin, particularly of the thighs and the upper arms following minor trauma, without associated bleeding into the muscles and joints and without abnormal bleeding in the mucous surfaces, are suggestive of purpura simplex or of "easy bruisability." Usually, the laboratory tests are negative in these patients with the possible exception of positive reaction to a tourniquet test and slightly prolonged bleeding time.

Many apparent hemorrhagic abnormalities are due to thrombotic phenomena. Skin lesions due to thrombi are likely to have a relatively pale center, surrounded by an area of ecchymosis, whereas a hemorrhage is likely to be most pronounced at the point of vascular rupture, with a fading out of extravasation peripherally. Hemorrhagic lesions on allergic basis are often elevated and may itch. Itching hemorrhagic lesions with linear petechiae along with scratch marks are evidences of allergic cause.

The age of the patient is important in determining the nature of the hemorrhagic lesions. In infants with hemophilia or hypoprothrombinemia, the superficial hemorrhages into the skin, which has excellent elastic tissue, may remain in tightly compartmentalized areas, producing nodules that resemble tumors. In older patients similar hemorrhages spread rapidly into the subcutaneous tissues and produce large, flat ecchymoses such as are seen in senile purpura.

Bleeding from more than one site is always of greater significance than hemorrhagic phenomena from a single area. Thus, an obstetrical patient who is in labor and is bleeding severely from the uterus is likely to have fibrinogenopenia if there is at the same time bleeding from the gums and extravasations of blood at the site of needle puncture wounds. A patient with bleeding from the uterus only, with no other signs of abnormality as revealed by physical examination and screening hemorrhagic studies, probably has a local lesion, rather than systemic disease.

The history and physical examination together with screening laboratory tests such as hematocrit and examination of a specimen of blood, including thrombocyte evaluation, are more reliable in predicting the tendency to bleed at the time of operation than are the bleeding time and coagulation time. If the patient who is scheduled for operation or for biopsy is bleeding from any site, or if there are symptoms or signs of abnormality in vascular or plasma coagulation components, preoperative hemorrhagic studies are indicated. On the other hand, routine preoperative hemorrhagic studies are not indicated in the case of patients who are not bleeding at the time, or who have a personal and family history negative for abnormal bleeding and no signs of abnormality at the time of physical examination.

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This concludes Part I of an article in two parts. Part II will be published next month.